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Self-management and skills acquisition in boys with haemophilia

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Abstract

Background There is an increasing prevalence of children/young people with long-term conditions (LTC) in the UK due to improvements in health-care management and delivery. These children are often involved, from an early age, in their own care and management; yet, there are little data to support how or when they develop the necessary skills and knowledge to become competent at this care.

Objective This study aimed to understand self-management of haemophilia, from a child's perspective, in the 21st century in the UK where intensive prophylactic therapy is given from early childhood.

Design A qualitative study using grounded theory to evaluate life-experiences of children and young people with haemophilia.

Setting and participants Thirty boys aged 4–16 with severe haemophilia treated at a single paediatric haemophilia care centre were interviewed at home or in a focus group.

Intervention/variables Multimethod qualitative research including age-appropriate research tools (draw and write, photo-elicitation and interviews) to facilitate data collection from children.

Results Boys develop self-management skills over time. They learn from health-care professionals, their parents and other family members with haemophilia.

Discussion Self-management skills (bleed recognition, self-infusion, self and medicines management, pain and risk management and conceptualizing preventative therapy) are developed through experiential learning and individualized education, and not through formalized expert patient programmes.

Conclusion The boys in this study have benefited from early prophylactic factor replacement therapy. They develop skills in haemophilia and self-management at a relatively young age and are experts in their own haemophilia care.

Introduction

Changing patterns of disease include an increase in the prevalence of long-term conditions (LTCs); Dowrick *et al.*[1] consider that part of the response to this increase should be the growth of patient empowerment through self-management. Jordan *et al.*[2] describe self-management as a 'patient's health literacy': their capacity to 'seek, understand and utilize health information to participate in decisions about health'. They cite five components to management: access (to information), knowledge, health education and empowerment, self-management and command (of the health-care system including confidence to use their initiative). Self-management in LTCs refers to 'daily activities that individuals undertake to keep illness under control, minimize its impact on physical health and psychological sequelae of the illness'.[3] Pritchard-Kennedy[4] argues that children's role as partners in care has often been confounded by parental dominance; thus, the 'self' in self-management in children is a combination of children and adult carers.[5]

This paper explores how boys with haemophilia develop expertise in self-management, becoming expert patients over time, through education and experience. Haemophilia reviews occur every 3–6 months during which the boys are encouraged to self-infuse and are asked questions about activity, treatment and genetics. This reflects Kirk *et al.*'s.[5] four-stage model of 'community, independence and confidence, knowledge and skills and engaging children/young people' in self-care. The boys in this study are the first generation to grow up with intense preventative treatment, (rather than treatment only after a bleed had occurred) recommended in the United Kingdom[6] with genetically engineered therapy that is known to be safe from bloodborne viruses,[7] as this has been shown to minimize disability and improve quality of life (QoL).[8]

Children are treated on home therapy programmes from early childhood,[9] We know little of how this experience impacts upon them, other than that this results in a lifestyle that is near to that of a child without haemophilia, where QoL scores, when assessed using haemophilia- and age-specific QoL assessment tools, are high.[10] This contrasts with prior research undertaken in the 1980's when many boys co-infected with bloodborne viruses, reported severe impact on life-experience.[11–13]

The study

The aim of the wider study, a multimethod study using age-appropriate research methods[14] from which this paper is derived, was to understand what living with haemophilia is like for boys in the 21st century. The study was undertaken between March and September 2009 at a paediatric haemophilia comprehensive care centre in the UK; ethical approval was granted by a local National Health Service Research Ethics Committee. Parental consent and patient assent were obtained from all participants at study entry. Forty-six boys aged 4–16 years were invited to participate; 16 refused for a variety of reasons including not being interested in the study or not wanting to talk about having haemophilia.

The methodology included photo-elicitation for boys aged 4–7,[15] draw and write techniques for 8- to 12-year-olds,[16] focus groups for those aged 13–16[17] and individual interviews for those teenagers who were unable to attend the focus groups. The younger cohorts of boys were given digital cameras, or 'art packs' 2 weeks before having individual interviews in their own homes, and were asked to either take photographs of their normal daily activities or to draw a picture or write a story about being themselves. These were used as icebreakers and/or aide memoire to enhance discussion about their lives in the preceding 2 weeks during the interviews, which lasted between 20 and 60 min and were semi-structured, asking questions about the effects of haemophilia on their lives, their friends, sporting activities, school, their family and things they like and do not like. They were tape-recorded, allowing the researcher to give full attention to the interviewee, what he was saying or doing at the time.

Open-ended prompt questions, such as 'can you tell me what it is like to (experience something?)' and 'other boys have said (something) has that ever happened to you?', were used to elicit their views of living with haemophilia. Further details of the methods used in this study are reported.[18]

The study was conducted using grounded theory, where each interview was analysed as it occurred; this enabled modifications and additions to the semi-structured questionnaires and continued until data saturation within an emerging framework occurred.[19] The recordings from the interviews and the focus group were professionally transcribed verbatim and listened to repeatedly by the lead researcher before analysis and manual coding. The content of the narratives could be grouped and coded into seven recurring themes, which were recognized by the research team as important aspects of modern day life with haemophilia. Six of these themes have been published elsewhere[20,21]; the seventh, development of self-management skills, is discussed here.

Results

Thirty boys with severe haemophilia A (n = 27) or B (n = 3) participated in the study.

Half the boys were the first in their family to be affected by haemophilia; the other half had a previous family history of haemophilia with an affected grandfather (n = 5), uncle (n = 3), older brother (n = 4) or multiple family members (n = 3). All of the boys were treated with prophylactic factor therapy from early childhood and were able to participate fully in normal family, school and peer-led activities.

What was striking through the interview analysis was that the boys were developing self-management skills: a concrete knowledge of bleeding mechanisms, risk avoidance and treatment, as well as self-infusion skills. All children with LTCs, particularly those with congenital disease, are involved in their own health care from early childhood. The level of involvement changes as children develop the physical and psychological means to cope with self-care and eventually self-management.[22] These self-management skills are learnt from parents and health-care workers, and often for boys with haemophilia, from other affected family members; there was evidence in our study of increasing self-management skills from a very early age.

Haemophilia management includes an ability to self-monitor, to be aware of subtle bodily fluctuations, using a range of resources, such as previous knowledge and experience[21,23] and knowing when to seek help from adults or health-care professionals. In boys with haemophilia, this includes knowledge of how their genetic mutation is expressed as symptoms[23] and how these should be assessed, treated, monitored and reported.

The major impacts of haemophilia on the life of the boys in this study are described in three themes: recognition of bleeds, self-infusion, and medicine and self-management, with an emphasis on sport. In the accounts below, pseudonyms have been used to protect respondent identities.

Recognition of bleeds

Unlike many LTCs, haemophilia is 'invisible' with few signs of illness unless there is an acute bleed. Even with total adherence to prophylactic treatment regimens, bleeds can occur as a result of trauma. Bleed recognition, described by boys as a bubbly, tingling feeling inside the joint and treatment strategies (when and how to treat and what dose to give) are integral parts of haemophilia self-management. Prophylaxis, which requires frequent infusions but offers the best bleed protection, is the treatment regimen most commonly used in boys with severe haemophilia:

> It's not really a big deal, it's just like as long as you do your injections when you are meant to you just crack on with it – a normal life, it's not like there is anything wrong with you, it's just simple. Haemophilia is something that only happens when you like, get hit or something, you'll bleed a bit more than the average person then you just need a bit more factor (Oliver aged 14)

Without early recognition of bleeding, or with recurrent bleeds into the same joint, boys with haemophilia might, in response to the pain of bleeding, withdraw from regular activities and social contacts; this may affect self-image[24] Paul aged 16 describes how bleeds impact on him:

> Sometimes just when I get a bleed it hurts loads so I take codeine at least twice but only because I think it knocks me out as well and puts me in kinda a place where well …… I just daydream or fall asleep and get rid of it [the pain] with luck. I think it puts you in that better state and if it takes a bit of the pain away then I think you are in a better state of mind, more chance of repairing it [the bleed] quicker

Paul describes the physical and psychological impact of bleeding, showing resilience to the pain and the social isolation that haemophilia can cause, which is more often seen in adults with haemophilia[25] However, he also shows that he is able to make appropriate treatment decisions, demonstrating skilled practice, competence, confidence and control, integrating haemophilia treatment options including analgesia as well as factor therapy into his personal management plan.[26]

Self-infusion

Peripheral venipuncture is a skill that many health-care professionals are unable to undertake, particularly in young children. In haemophilia management, parents learn this skill to facilitate home treatment of their sons,[27] who learn to self-infuse in late childhood or early adolescence.[28] Even though this is a routine treatment for boys, becoming self-sufficient at infusion is complex, most boys can do this at least some of the time by the age of twelve. To infuse into one arm makes the use of that arm for anything other than venipuncture impossible; thus, self-infusion becomes a one-handed technique. They need to apply a tourniquet, find a vein, puncture the vein, release the tourniquet, infuse the factor, and most difficult of all remove the needle and apply pressure to the puncture site at the same time. This sometimes occurs in the face of a bleed in the infusing arm, further reducing mobility. Many boys become ambidextrous at this skill; this gives them more choice of venipuncture sites and more ability to repeatedly infuse.

> At first I felt quite scared, when I done it [sic] I thought to myself 'ok this is it one, two, three' and then I put it in and it did not really hurt that much and then I was really happy. Well I was scared of if I went wrong because it's quite scary sticking a needle into yourself. Before when it went wrong it didn't really hurt but I knew I went wrong because it started to balloon [extravasate] and so we [Tom and his mum] took it out and then I tried again in my right arm with my left hand and that went wrong as well because I am right handed and it was quite hard with my left hand (Tom aged 12).

Starting to become a self-infuser often raises anxiety around what will happen if the infusion goes wrong – in the past, a parent will have been the main infuser and will have had the necessary skills to 'fix' the infusion if a vein is missed or if extravasation occurs as described by Tom above and 10-year-old Jonathan:

> I am finding it a little bit hard, because I am frightened that if I do it wrong then I might die, because if I do it wrong maybe the vein will puff up into a lump and pop. It would hurt. Sometimes I stick the needle in, that feels weird cause one hand is actually pushing down and one hand has actually got it in, and it's like pins and needles [due to tourniquet] while I'm doing it. It just feels really weird.

Tom and Jonathan show how becoming competent at self-infusion, itself a process of skill and expertise developed through trial and error as well as education and support, enables them to start on the path to becoming a patient with expert skills for current haemophilia care. For boys with haemophilia, self-management includes an understanding of not just how, but when to treat themselves.

Medicine and self-management

Treatment with replacement therapy, both prophylactically and 'on demand' when bleeds occur, is anchored in everyday life for boys

with haemophilia. Routine, such as having treatment at the same time every day, is essential to remain concordant with treatment.[29] The ability to control symptoms and the severity of bleeding achieve some normality and control in daily life.[30] Ingadottir *et al.*[31] discuss how, in children with diabetes, self-knowledge is key for integrating treatment regimens and achieving good QoL. They describe a concept of 'body-listening' – knowing when to treat and developing self-management strategies which suit lifestyle, belief patterns and personal priorities. However, in haemophilia, 'body-listening' (taking a 'wait and see' approach) or missing treatment will result in the rapid onset of bleeds which are painful and ultimately result in joint damage[32] and increased pain as shown by 10-year-old Will:

> ankle bleeds I can always tell when they are going to hurt and when they are not …… it feels a bit stiff and then later can be oh it's just really really hurting.

Understanding how to manage treatment, including storage, reconstitution and infusion[33] how to integrate this into daily life and how to develop sustainable routines, is a key part of medicine management.[29] Motivated adolescents are more likely to demonstrate good adherence[34] as they then have less restriction placed on lifestyle choices. Oliver aged 14, a keen golfer, showed a good understanding of the rationale for a daily treatment regimen, including the concept of factor half life, where factor degenerates and does not accumulate in the body, thus requiring regular infusion:

> Because then it [the factor level] will be on one consistent level because at the moment it's like if you had a graph it would be going up down, up down, up down so if you have it every day it would be pretty straight. Because like if you are on a low, and you had an injury, you would be even more smashed up but if, because it's all on one level its like you won't get [hurt] as much.

Adolescents handle illness and related situations, by 'focusing on disease and its treatment for self protective reasons',[35, p. 283]. Hinds,[35, p. 283] shows that adolescents shift from 'wellness-in-the-foreground to illness-in-the-foreground' when they are in situations of 'threat, transition, suffering, pain or coping with physical or psychological limitations of disease or treatment'. For boys with haemophilia, life without it is unimaginable[25,36] and it is integral to who they are as individuals:

> [I] suppose it just becomes part of you doesn't it; it's what you think about. I find it's just life so I kind of just make it part of my life so just do certain things to ensure that I keep myself safe and kind of take it in your [sic] stride. (Peter aged 14)

The concept of 'this is it/normal for me' is described by Atkin & Ahmad[37] who show that adolescents with haemoglobinopathies emphasize the importance of 'just getting on with life' and 'limiting the consequences of their illness on their day-to-day lives' by using normalization strategies to maintain 'normal' lives and identities. For a majority of boys, 'normalization' includes the ability to participate in sport. Koiter *et al.*[38] state that choices around specific sporting activity, often considered 'dangerous' for boys with haemophilia, must be based on adaptation to the haemophilia diagnosis and treatment regimen. Adolescent boys are aware of potential risks that sporting activity may pose and may participate with caution. They may adjust their normal medication routine, gaining control over their haemophilia, to be able to participate in sport, further demonstrating medicine management skills[39] as well as risk-taking (or avoiding) strategies.

Christensen & Mikkelsen[40] suggest that children strategize risk management in everyday life. They judge and balance the chances and risks they encounter, learning to control and steer through these, distinguishing between negative risk-taking and positive risk management. Risk-taking and pushing at boundaries is an activity especially valued amongst boys, who generally engage in more physical activity, such as contact sports, where injury is an accepted risk of participation, as they 'prove themselves'.[41] For boys with haemophilia, the choice of sporting activity may be affected by the diagnosis[39] and the treatment as well as by child/family expectations.

Contact sports are undertaken with extreme caution[32]; many younger boys participate but choose to adapt their activity as they (and their opponents) become bigger. Mark (aged 16) described how he chose to manage the risk of sporting participation by adapting his sporting choice to one with less contact:

> I used to do football and golf, but not now – no sport like that for me now or I'll be in bed for a week with a bad back – so just swimming or cycling.

Mark treats himself with prophylaxis punctiliously but has decided that this positive risk management is still outweighed by the negative risk-taking of participating in contact sport now that he and his opponents are bigger. Haemophilia demands adjustments to avoid risk, lifestyle disruption and uncertainty associated with trauma-related bleeding.[30] For the majority of boys, the risk is manageable and causes no loss or biographical disruption to themselves. However, global limitations in haemophilia care can cause frustration; George, a 16-year-old polo player, expresses feelings of biographical disruption and loss[42] because of haemophilia:

> It [haemophilia] doesn't really stop me from doing anything except living in a load of other countries or spending a long time like a gap [year] in Argentina, which is a bit annoying.

The risks for George of a gap year as a gaucho, so far from home, are deemed to outweigh the benefits, something with which he has, with reluctance, come to terms.

Discussion

Like other adolescents with LTCs, boys with haemophilia are responsible for their own health, making daily changes in lifestyle and treatment choices, although their mothers act as 'alert assistants'.[43] These include healthy eating, participation in exercise, getting enough sleep and making other health choices such as not smoking. Taylor *et al.*[44] argues that children and young people with LTCs strive for 'normalcy'; the focus of their care should therefore be on making and taking healthy choices which result in wellness rather than illness. In haemophilia, wellness is achieved through individualized treatment, self-care and an understanding of medicine management which reduces bleed frequency.

Unlike many other LTCs, haemophilia, if untreated, leads to a rapid onset of bleeding which requires the same treatment. This rapid feedback of poor self-management resulting in rapid symptoms is unlike most other LTCs where missing or stopping treatment rarely results in visible changes to wellness. Usually in boys with haemophilia, wellness is in the foreground of their daily experience. Paterson[45] shows that illness moves to the foreground of people's worlds when they have an acute crisis, which forces them to attend to the illness. In haemophilia, the event would be a joint bleed, with its associated decreased range of movement, pain and the need for further factor therapy. The paradox of this model is that to keep wellness in the foreground, disease management must also be foremost; 'the illness requires attention in order not to pay attention to it',[45, p. 24], and the body becomes something to which things are done, in this instance by regular self-infusions. The routine of this treatment becomes immersed in the everyday normality of haemophilia care such that haemophilia *per se* can be forgotten.[29] There is therefore little impact on day-to-day activity as long as treatment is adhered to.[21]

The core category of this study is how young people with haemophilia have developed self-management skills. Young people with haemophilia are an 'untapped resource of expertise, that often know more about their particular condition and its management than their doctors, and this knowledge may assist treatment',[36, p32]. However, adolescents with LTCs need to demonstrate more 'refined social skills than their healthy peers, since they have to be able to accommodate treatment requirements and social demands',[40, p. 26] into their everyday lives. Daily life needs to be planned to reduce the risk of bleeding, so that haemophilia does not come to the foreground, removing wellness from its prime position. This may encompass

adapting the medical treatment regimen to fit within other priorities of daily life, such as school and sport so that daily routines are contextualized within both illness sequelae and personal experience that is reflective of (and sometimes conflicting with) lived events, experiences and perspectives.[22]

An expert patient is one who has the necessary skills to be a competent individual. These include knowledge, clinical judgement, self-efficacy, physical skills, persistence, paying attention to tasks and integrating these into daily management.[26] The boys in this study identify with others on the same 'illness trajectory'[23] where they recognize the consequences of haemophilia in other family members. Mark tells how his older brother who also has haemophilia '*keeps an eye out for me too'* by recognizing subtle signs of bleeding which their non-haemophilic brother would not necessarily notice:

> we kind of ….you know we have this kinda relation thing where …….. you know you walk along and you are hobbling a bit perhaps and he [his brother with haemophilia] goes 'oh you've got a bad ankle'. He would always notice.. and tell me to get a needle [factor infusion]

These boys have access to treatment which their older haemophilic family members did not have and are able to tailor their own treatment around sporting or other risk activities. All children make mistakes in their judgements; when they do this, they often find solutions to the mistake in their own ways.[40] This is demonstrated by 16-year-old Mark who had adapted his sporting activities to those which involve less contact as he recognizes that contact sports now have more risk than benefit for him as an individual. The independence gained by young people with LTCs as they grow older allows them more autonomy over their treatment, making it easier for them to appear 'normal' to others.[36] For Mark being able to infuse before cycle races means that several people in his cycling club are unaware that he has haemophilia (data not shown); thus, he is not defined by haemophilia but as a fellow cyclist.

Strengths and limitations

The strengths of this study include the use of age-appropriate research methods and the provision of insights into children's views of living with haemophilia where prophylaxis enables near normal lifestyles. The study is limited as it was undertaken in a single paediatric haemophilia comprehensive care centre in the UK; thus, the results of this study cannot be extrapolated to children and young people with haemophilia in other centres or those treated with different therapeutic regimens. Further work is necessary to confirm these findings with others who self-manage haemophilia through home treatment programmes. However, this study confirms the need to further discover the experiences and impact of haemophilia on children and young people, to continue to shape how we support them in developing self-management strategies.

Conclusion

The boys in this study are amongst the first generation in the UK to be offered safe and effective treatment, enabling them to lead more 'normal' lives than any person with haemophilia before them. They received prophylactic therapy from an early age and consequently do not have joint damage which has historically, limited mobility and sporting activity. Thus, they have grown up describing themselves as they see their peers, as normal young men who participate fully in day-to-day life. However, if they fail to treat themselves, they develop bleeds which are painful and debilitating, reminding them that they have haemophilia.

Haemophilia knowledge and education begins in early childhood, with basic recognition of bleeding and infusion skills being taught to young boys by health-care professionals. Other education and learning occurs on an 'ad-hoc', individualized basis with most boys learning through personal experience, this is described by Kirk *et al.*[5] as 'individualized self-care projects' which 'maintain involvement'. Some boys have other, usually older,

affected family members, from whom knowledge and skills can also be gained. All boys with haemophilia are experts, and as Petersen[30] states, they are probably the best haemophilia experts that there are.

There are no haemophilia-specific training courses for children or adolescents with haemophilia, and many boys have declined the opportunity to participate in programmes aimed specifically at adolescents with LTCs. For some, this is because they do not want to be defined by haemophilia. The boys in this study have developed expert patient skills. They are without doubt expert patients, from whom younger boys with haemophilia may gain peer support in relation to performing self-care (self-infusion techniques); these boys do not have the benefit of a generation before them who have had access to the same care and treatment; they are the first generation of experts with haemophilia who perceive themselves to have normal lives and are a great resource for haemophilia care in the future.

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Conflict of interest

None.

References

1 Dowrick C, Dixon-Woods M, Holman H, Weinman J. What is chronic illness? *Chronic Illness*, 2005; **1**: 1–6.

2 Jordan J, Briggs M, Brand A, Osborne R. Enhancing patient engagement in chronic disease self-management support initiatives in Australia: the need for an integrated approach. *Medical Journal of Australia*, 2008; **189** (10 Suppl): S9–S13.

3 Jones M, MacGillivray S, Kroll T, Reza Zohoor A, Connaghan J. A thematic analysis of the conceptualisation of self-care, self-management, and self-management support in the long-term conditions management literature. *Journal of Nursing and Healthcare of Chronic Illness*, 2011; **3**: 174–185.

4 Pritchard Kennedy A. Systematic ethnography of school-age children with bleeding disorders and other chronic illnesses. *Child: Care, Health and Development*, 2012; **38**: 863–869.

5 Kirk S, Beatty S, Callery P, Milnes L, Pryjmanchuk S. Perceptions of effective self-care support for children and young people with long-term conditions. *Journal of Clinical Nursing*, 2013; **39**: 305–324.

6 Richards M, Williams M, Chalmers E *et al.*, Paediatric Working Party of the United Kingdom Haemophilia Doctors' Organisation A United Kingdom Haemophilia Centre Doctors' Organization guideline approved by the British Committee for Standards in Haematology: guideline on the use of prophylactic factor VIII concentrate in children and adults with severe haemophilia A. *British Journal of Haematology*, 2010; **149**: 498–507.

7 Keeling D, Tait C, Makris M. Guideline on the selection and use of therapeutic products to treat haemophilia and other hereditary bleeding disorders. A United Kingdom Haemophilia Centre Doctors' Organisation (UKHCDO) guideline approved by the British Committee for Standards in haematology. *Haemophilia*, 2008; **14**: 671–684.

8 Liesner R, Khair K, Hann IM. The impact of prophylaxis on children with severe haemophilia. *British Journal of Haematology*, 1996; **92**: 937–938.

9 Khair K. Managing haemophilia at home. *British Journal of Home Healthcare*, 2006; **1**: 10–11.

10 Gringeri A, von Makensen S, Auserwald G *et al.*, for the Haemo-QoL study Health status and health related quality of life of children with haemophilia from six west European countries. *Haemophilia*, 2004; **10**: 26–33.

11 Goldman E, Miller R, Lee C. Counselling HIV positive haemophilic men who wish to have children. *British Medical Journal*, 1992; **304**: 829–830.

12 Goldman E, Miller R, Lee C. A family with HIV and haemophilia. *AIDS Care*, 1993a; **5**: 79–85.

13 Goldman E, Lee C, Miller R, Kernoff P, Morris-Smith J, Taylor B. Children of HIV positive haemophilic men. *Archives of Disease in Childhood*, 1993b; **68**: 133–134.

14 Coad J, Plumridge G, Metcalfe A. Involving children and young people in the development of art-based research tools. *Nurse Researcher*, 2009; **16**: 56–64.

15 Clark C. The autodriven interview. A photographic viewfinder into children's experience. *Visual Sociology*, 1999; **14**: 39–50.
16 Horstmann M, Aldiss S, Richardson A *et al*. Methodological issues when using the draw and write technique with children aged 6–12 years. *Qualitative Health Research*, 2008; **18**: 1001–1011.
17 Gibson F. Conducting focus groups with children and young people; strategies for success. *Journal of Nursing Research*, 2007; **12**: 473–483.
18 Khair K, Collier C, Meerabeau E, Gibson F. Multi-methodology research with boys with haemophilia. *Nursing Researcher* 2013; in press.
19 Charmaz K (2006) *Constructing Grounded Theory. A Practical Guide Through Qualitative Analysis*. London: Sage.
20 Khair K, Gibson F, Meerabeau L. 'Just an unfortunate coincidence': children's understanding of haemophilia genetics and inheritance. *Haemophilia*, 2011; **17**: 47–75.
21 Khair K, Gibson F, Meerabeau L. The benefits of prophylaxis: views of adolescents with severe haemophilia. *Haemophilia*, 2012; **18**: e286–e289.
22 Nicholas DB, Picone G, Selkirk EK. The lived experiences of children and adolescents with end-stage renal disease. *Qualitative Health Research*, 2011; **21**: 162–173.
23 Giarelli E, Bernhardt BA, Mack R, Pyeritz RE. Adolescents transition to self-management of a chronic genetic disorder. *Qualitative Health Research*, 2008; **18**: 441–456.
24 Hegeman AK, Van Genderen FR, Meijer S, Van Den Briel MM, Tamminga RYJ, Van Weert E. Perceived competence in children and adolescents with haemophilia: an explorative study. *Haemophilia*, 2011; **17**: 81–89.
25 Beeton K, Neal D, Lee C. An exploration of health-related quality of life in adults with haemophilia –a qualitative perspective. *Haemophilia*, 2005; **11**: 123–132.
26 Redman BK. Accountability for patient self-management of chronic conditions; ethical analysis and a proposal. *Chronic Illness*, 2007; **3**: 88–95.
27 Vidler V. Teaching parents advanced clinical skills. *Haemophilia*, 1999; **5**: 349–353.
28 Lindvall K, Colstrup L, Wolter IM *et al*. Compliance with treatment and understanding of own disease in patients with severe and moderate haemophilia. *Haemophilia*, 2006; **12**: 47–51.
29 Haslbeck JW, Schaffer D. Routines in medication management: the perspective of people with chronic conditions. *Chronic Illness*, 2009; **5**: 184–196.
30 Petersen A. The best experts: the narratives of those who have a genetic condition. *Social Science and Medicine*, 2006; **63**: 32–42.
31 Ingadottir B, Halldorsottir S. To discipline a 'dog': the essential structure of mastering diabetes. *Qualitative Health Research*, 2008; **18**: 606–619.
32 Park J. 'The worst hassle is you can't play rugby': Haemophilia and masculinity in New Zealand. *Current Anthropology*, 2000; **41**: 443–452.
33 Khair K. Children's preferences of transfer devices for reconstitution of factors VIII and IX for the treatment of haemophilia. *Haemophilia*, 2009; **15**: 159–167.
34 Kyngas H. Predictors of good adherence of adolescents with diabetes. *Chronic Illness*, 2007; **3**: 20–28.
35 Hinds P. Shifting perspectives. *Oncology Nursing Forum*, 2004; **31**: 281–287.
36 Williams B, Corlett J, Dowell J, Coyle J, Mukopadhyay S. 'I've never not had it so I don't really know what its like not to': nondifference and biographical disruption among children and young people with cystic fibrosis. *Qualitative Health Research*, 2009; **19**: 1443–1455.
37 Atkin K, Ahmad W. Living a 'normal' life: young people coping with thalassaemia major or sickle cell disorder. *Social Science and Medicine*, 2001; **53**: 615–626.
38 Koiter J, Van Genderen FR, Brons PPT, Nijhuis-Van Der Sanden MWG. Participation and risk-taking behaviour in sports in children with haemophilia. *Haemophilia*, 2009; **15**: 686–694.
39 Conrad P. The meaning of medications: another look at compliance. *Social Science and Medicine*, 1985; **20**: 29–37.
40 Christensen P, Mikkelsen MR. Jumping off and being careful: children's strategies of risk management in everyday life. *Sociology of Health & Illness*, 2007; **30**: 112–130.
41 Green J. Risk and the construction of social identity: children's talk about accidents. *Sociology of Health & Illness*, 1997; **19**: 457–479.
42 Williams S. Chronic illness as a biographical disruption or biographical disruption as chronic illness? Reflections on a core concept *Sociology of Health & Illness*, 2000; **22**: 4–67.
43 Williams C. Alert assistants in managing chronic illness: the case of mothers and teenage sons. *Sociology of Health & Illness*, 2000; **22**: 254–272.
44 Taylor R, Gibson F, Franck L. The experience of living with a chronic illness during adolescence: a critical review of the literature. *Journal of Clinical Nursing*, 2008; **17**: 3083–3091.
45 Paterson B. The shifting perspectives model of chronic illness. *Journal of Nursing Scholarship*, 2001; **33**: 21–26.